Diabetic Neuroarthropathy: Report of Four Cases

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ABSTRACT

Diabetic neuroarthropathy was observed in four patients; these are the first cases of this nature reported in the Canadian medical literature. The criteria for this diagnosis included: (1) long-standing diabetes; (2) arthropathy, most frequently involving the foot, which shows deformity, shortening and ulceration without evidence of infection or peripheral circulatory failure; (3) abolition or diminution of pain on weight-bearing; (4) diabetic peripheral neuropathy with impaired sense of position or vibration and weak or absent deep tendon reflexes. Radiographic findings were similar to those in patients with Charcot's arthropathy from any cause.

Tabes dorsalis, leprosy, syringomyelia, myelodysplasia and the arthropathies of corticosteroid therapy were ruled out in these cases. In addition to conventional medical therapy the patients were treated by means of walking-casts for several months.

Diabetic neuroarthropathy is probably more common than the medical literature would indicate. Diminished sensation in the lower limbs in diabetics of long standing appears to be the major factor contributing to this disorder.

SOMMAIRE

Des neuroarthropathies diabétiques ont été observées chez quatre malades et ces cas ont été les premiers à être rapportés dans la littérature médicale Canadienne. Les critères pour ce diagnostic inclurent 1) un diabète de longue évolution, 2) une localisation très fréquente au pied qui s'est déformé, raccourci, affaissé, ulcéré sans signe d'infection et d'insuffisance dans la circulation périphérique, 3) une absence totale ou une diminution de la douleur à la marche sur le pied déformé, 4) une neuropathie diabétique périphérique: réduction ou abolition du sens de position ou des vibrations avec hypo- ou aréflexie tendineuse. Les radiographies furent superposables à celles d'une arthropathie de Charcot.

Pour arriver à prouver ces neuroarthropathies diabétiques, nous avons éliminé le tabès, la lèpre, la syringomyélie, la myélodysplasie et les arthropathies consécutives à la corticostéroïdothérapie.

Les neuroarthropathies diabétiques sont, à notre avis, plus fréquentes que semble l'indiquer la littérature médicale. La diminution de la sensibilité aux membres inférieurs chez les diabétiques de longue évolution nous paraît être la cause principale des cas de neuroarthropathies diabétiques.

SINCE the discovery of insulin, late complications of diabetes, such as neuropathy, retinopathy and nephropathy, have been observed with increasing frequency.[39, 42, 54] Within the past few decades, Charcot's joints and painless neuropathic ulcers of the lower limbs, although frequent in patients with tabes dorsalis and syringomyelia, have appeared more often as complications of diabetes where control of the disease has been inadequate.[14, 44]

CASE REPORTS

CASE 1.—R.C., a 53-year-old man, was obese and had a slight elevation of blood pressure; his diabetes was discovered about two years previously. Because of the complaint of intense thirst present since 1950, it is assumed that his diabetes had been present for many years before 1961. A low-calorie diet (carbohydrate 132 g., fat 50 g., protein 58 g.) and NPH insulin from 18 to 26 units daily were administered. With this treatment the patient's urine was sugar-free except on one occasion. His blood sugar level varied from 118 to 160 mg. %. A year previously he had stepped on a nail with his left foot, and a plantar fistula had formed between the first and second metatarsal heads. This fistula drained for about six months and then dried up. Two months later, a new area of fistulous necrosis appeared under the third metatarsal head. During that period, he went about without obvious pain when walking. This patient was considered to have mild diabetes; administration of insulin appeared to result in more rapid healing of his wounds.

Clinical examination revealed a slight shortening of the first, second and third rays of the foot (Fig. 1) and a local area of necrosis under the third metatarsal head that had a tendency to crust. A funnel-shaped depression, which was not discharging, was noted between the first and second metatarsal heads. A slightly indurated edema of the whole forefoot was present. The dorsal aspect of the forefoot, overlying the metatarso-phalangeal joint of the third toe, was slightly reddened. The skin of the foot was dry and scaling. All of the toes were in a moderate claw position with plantar protrusion of the metatarsal heads.

A joint study by the Diabetic and Nutritional, and Orthopedic Services of the Hôpital du Sacré-Cœur de Montréal.
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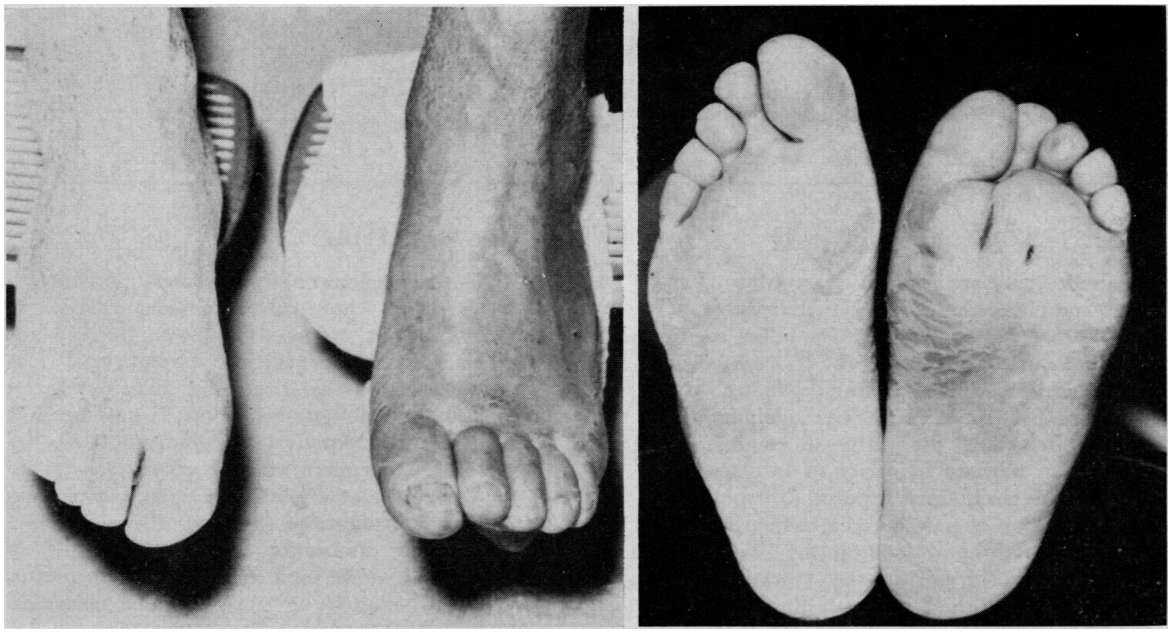

Fig. 1.—Case 1: Shortening of the left foot with localized necrosis beneath the third metatarsal head and a funnel-shaped cavity between the first and second metatarsal heads are seen. The skin is dry and peeling.

When the foot was raised, the circulation appeared to be normal; when lowered, a noticeable capillary congestion was present. The femoral, popliteal and posterior tibial pulsations were of good quality, and the pedal pulse was faintly perceived.

The patient never complained of paresthesias in the upper or lower limbs and had not noticed any decrease of peripheral sensitivity. The neurological examination revealed that the deep tendon reflexes were weak in the upper limbs but could be obtained after reinforcement in the lower limbs. There was no decrease of muscular strength in the lower limbs, nor any noticeable muscular atrophy. Position sense was absent in the toes and there was considerable decrease of vibration sense, most noticeable in the great toes and to a lesser degree at the malleoli. Light touch was preserved in the lower limbs but perception of pin-prick was distinctly reduced in the distal two-thirds of the feet. There was no Romberg sign. A pseudo Argyll Robertson pupil was noted; in fact, the pupils were slightly oval, and the reaction to light and accommodation was slower than normal.

Examination of the fundi revealed arteriovenous nicking with "copper-wire" arteries. There were many small punctate hemorrhages with a few clusters, indicating a diabetic and arteriosclerotic retinopathy. A radiograph of the foot showed destruction by osteolysis of the metatarsal heads and bases of the proximal phalanges of the first, second and third metatarsophalangeal joints (Fig. 2). A proliferative callus was noted on the first metatarsal. A radiograph of the other foot was negative, as was that of the wrists and hands, thus eliminating the possibility of syringomyelia where destruction usually spreads progressively to the hands and feet. Radiography of the lumbosacral spine was also normal, eliminating rachischisis or spina bifida. These myelodysplasias cause plantar ulcers with osteoarticular destructions similar to those seen in patients with Charcot's tabetic foot.

The clinical picture allowed the elimination of leprous arthropathy. Likewise, syphilis was eliminated by a negative *Treponema pallidum* immobilization test (T.P.I.), by the absence of Romberg sign and tabetic pains, and above all by negative serological reactions of the blood and cerebrospinal fluid. Infectious arthritis was eliminated by the absence of local heat, a normal white blood count, and negative cultures of the fistula.

During the first two weeks of hospitalization, the patient was treated by bed rest and exposure of the foot without a dressing. A progressive healing of the plantar sore occurred; it dried, crusted and finally epithelialized. A walking-cast was then applied for an intended period of four to six months, during which time control radiographs were to be taken every two months without the cast.

CASE 2.—Mrs. A.M., a 58-year-old obese (230 lb.) diabetic patient, had been taking insulin since 1953.

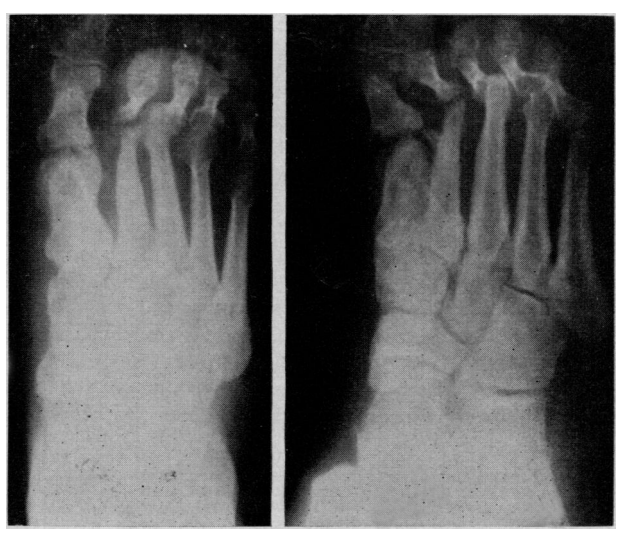

Fig. 2.—Case 1: Osteolysis of the first, second and third metatarso-phalangeal joints is apparent. Proliferative callus is seen along the first metatarsal.

She was admitted because of a suppurative lesion on the fourth toe of the right foot and abnormal mobility of this digit. She gave no history of trauma, except for the abnormal strain of her excessive weight, and she felt no pain on walking. She had borne five children, the largest weighing 11½ lb. at birth. She had had chronic cervical adenitis in her youth which left a residual scar. Her father and one of her sisters were also diabetic. The functional inquiry revealed nothing remarkable. She had a good appetite and digestion. There was no nocturnal diarrhea, but a slight tendency to constipation.

On physical examination, in addition to her obesity a slight exophthalmos with divergent strabismus was noted and also a corneal opacity of the right eye, as well as slowly reacting pupils. The thyroid gland was normal. Blood pressure was 150/80 mm. Hg. The abdomen was huge but soft. Examination of the limbs showed the presence of patellar reflexes but absence of the Achilles reflexes. There was no decrease of muscular strength in the lower limbs, but absence of position sense in the toes. Light touch perception was diminished in the right lower limb, with decreased perception of pin-prick in the foot.

The patient was seen in consultation by an ophthalmologist. Owing to the external deviation of the right eye since the age of 4, the vision was very poor. Scarred central and paracentral corneal opacity was present. The pupils were equal, of normal diameter, and responded with very slow and weak reaction to light. The right eye distinguished hand movements, while in the left the sight was 20/400. The ophthalmologist did not interpret this as definite evidence of an Argyll Robertson pupil, since sluggish pupils can occur in diabetes. The patient was also examined by a consultant in vascular surgery who noted that the right foot was warm and of good colour, and that a palpable pulse was present in the posterior tibial artery; in fact, the peripheral circulation was still satisfactory. Oscillometry suggested limitation of peripheral circulation on the right, but it was adequate on the left.

The orthopedic surgeon noted that the right foot was shortened and that the medial aspect of the tarsal region bulged outward. The fourth toe was shortened, deformed, ulcerated and almost flail-like (Fig. 3).

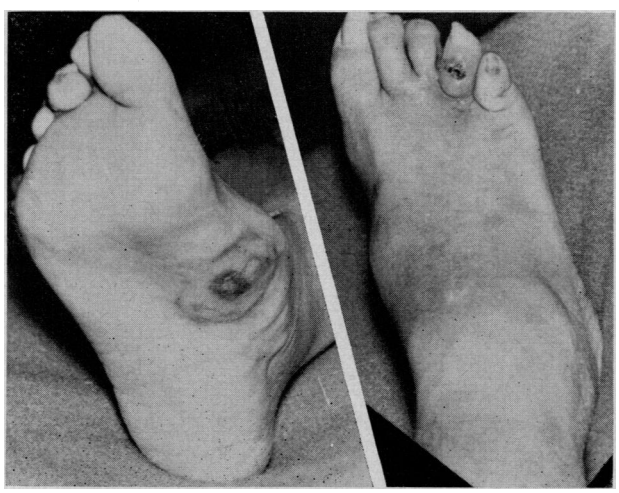

Fig. 3.—Case 2: An ulcerated and oozing bulging is present on the medial aspect of the right tarsus. The fourth toe is deformed, flail-like and ulcerated.

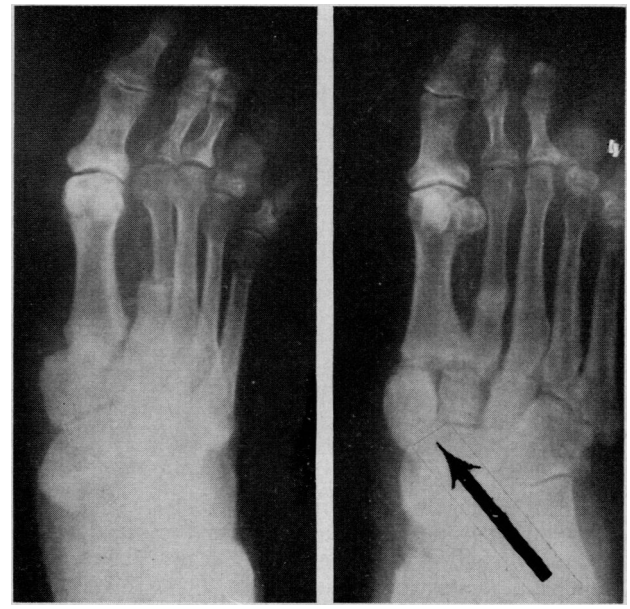

Fig. 4.—Case 2: Medial subluxation of the cuneiforms and destruction of the fourth and fifth toes.

The bulge on the right instep was ulcerated and oozing. Radiography showed an old march fracture

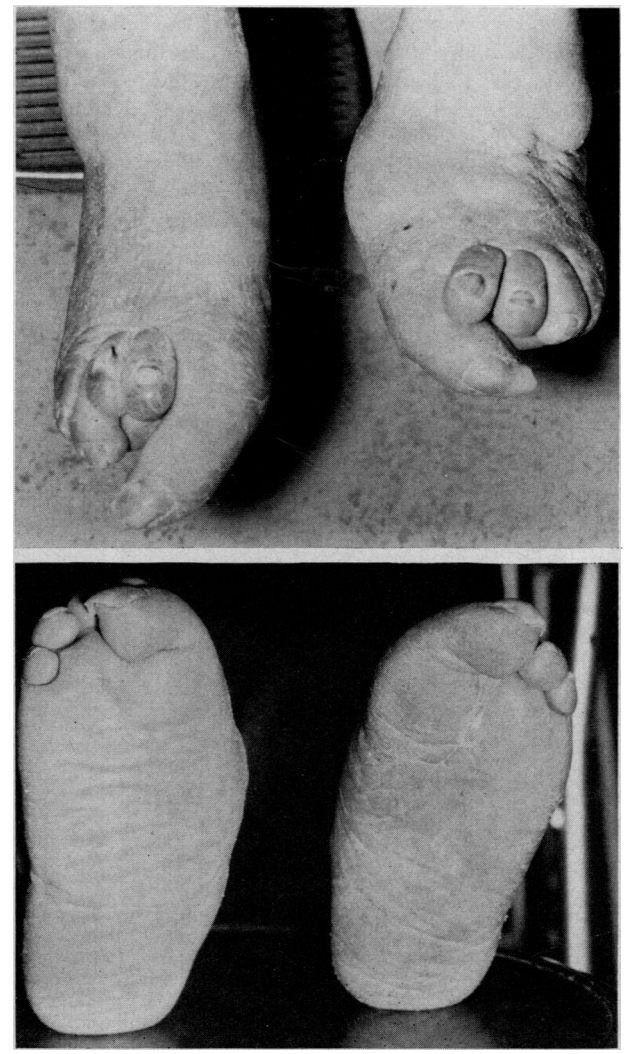

Fig. 5.—Case 3: Charcot's cube-shaped feet.

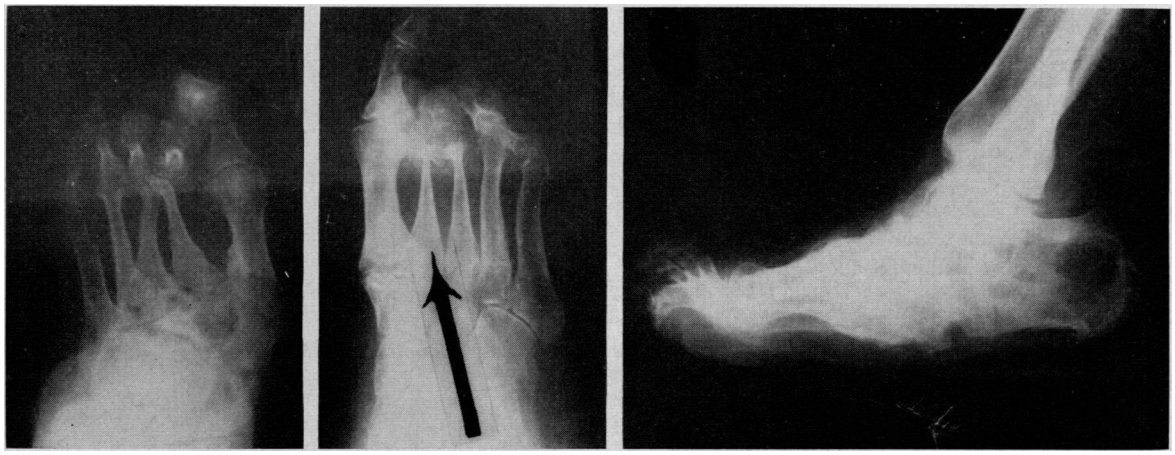

Fig. 6.—Case 3: Metatarsal tapering like a "sucked candy-stick".

at the middle and proximal thirds of the second right metatarsal shaft. There was a subluxation in the tarso-metatarsal joints which showed an incongruity by inward displacement of the first cuneiform, causing the bulge of the instep (Fig. 4). The phalanges of the fourth and fifth toes were partially destroyed.

Two weeks' bed rest healed the skin lesions completely, and a walking-cast was applied. The latter was replaced by a molded shoe a few months later.

CASE 3.—Mrs. O.G., 78 years of age and a known diabetic for 15 years, was being treated with a diet and oral hypoglycemic agents. She was hospitalized after a fall at home which caused a fracture of the right patella without displacement. The patient was very deaf and had been almost blind for the past five or six years owing to bilateral cataracts. Her memory was unreliable. She bore three children who are still living. At the age of 50, she underwent a hysterectomy with appendectomy and some type of surgery of the urinary bladder. Eight years previously, the patient's left hip was nailed and she also broke her left ankle. Finally, she had fractured her left wrist 20 years ago. Three of her sisters also had diabetes.

Physical examination showed a woman of her stated age, very deaf, with bilateral cataracts which allowed almost no vision. The cardiac rhythm was regular and she had a blood pressure of 170/85 mm. Hg. The abdomen was soft on palpation and there was no enlargement of the liver or spleen. Marked and painless deformities of both feet were present. The feet were shortened, splayed and cubical, with marked convexity of the medial border (Fig. 5). The toes were also misshapen: the great toe was in a valgus deformation under the second and third claw-toes. No sores or ulcerations were present on the weight-bearing surfaces of the feet. A complete biochemical and radiological examination was carried out, ophthalmological and neurological consultations were obtained and a lumbar puncture was done.

Her visual acuity was 20/300. Both corneas were normal and the pupils were equal, reacting to light and accommodation. Ocular mobility and tension were

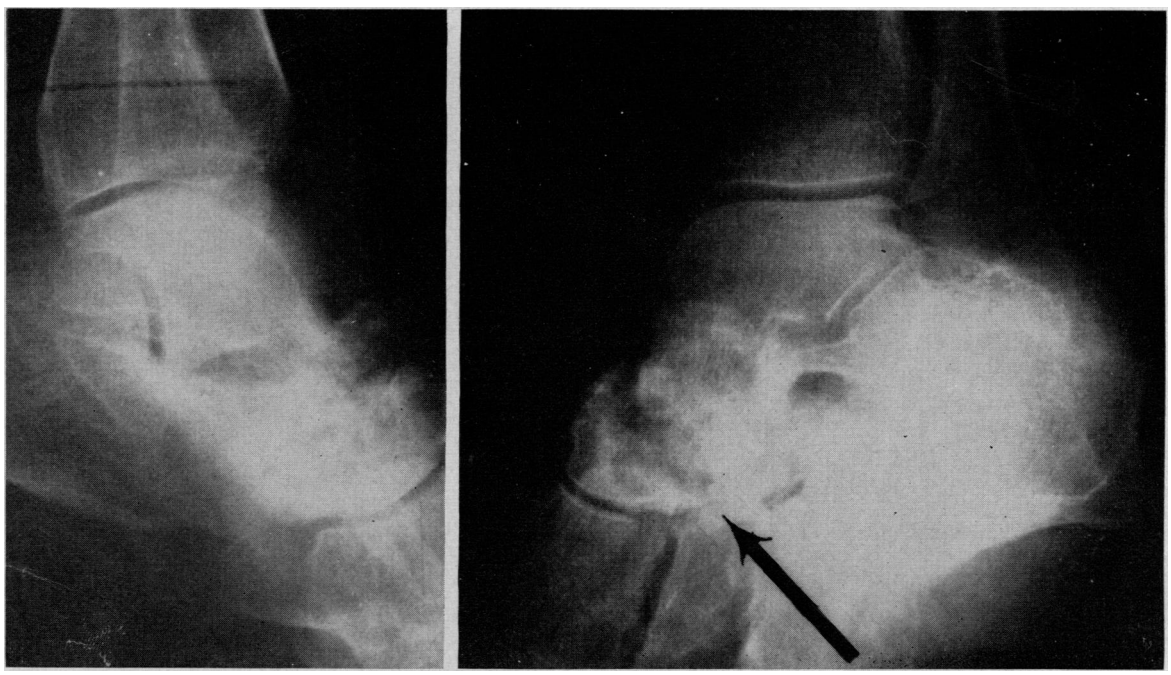

Fig. 7.—Case 4: Painless destruction of the talo-scaphoid joint.

TABLE I.—LABORATORY RESULTS IN THE FOUR CASES

	R.C., male, 53	*A.M., female, 58*	*O.G., female, 78*	*L.C., female, 35*
E.C.G.	Normal	Normal	Normal	Ventricular hypertrophy Ischemia Pericarditis
Fasting blood sugar	Between 110 and 180	Between 80 and 200	Between 150 and 151	Between 40 and 610 mg.%
Glucosuria	Negative	Negative	Between 0 and 13.2	Between 0 and 53 g./l.
Hemogram	Hb. 13.6 Ht. 41 W.B.C. 5300 Polym. 65 Baso. 1 Lympho. 32 Mono. 2 Eosin. 0	10.6 30.0 R.B.C. 3,740,000	11.6 34.0 W.B.C. 11,700 59 0 34 6 1	7.6 g.% 23.0% W.B.C. 11,300 per c.mm. 87% 0 10 2 1
Sedimentation rate	25	34	29	26 mm. per hr.
Blood urea nitrogen	17	32	19	90 and 156 mg.%
Bacteriology	Negative	*Staphylococcus pyogenes*	Negative	Negative
Cerebrospinal fluid	Proteins 36.8 Cells—absent W.R. negative	45.5 Absent Negative	38.6 Absent Negative	85 mg.% Absent Negative
V.D.R.L.	Negative	Negative	Negative	Negative
T.P.I.T.	Negative	Not done	Not done	Not done

normal. Immature cataracts, present in the vitreous, obscured retinal details. Deep tendon reflexes were absent but superficial and deep sensation was not disturbed. Position sense was impaired in both feet, and hypoesthesia was noted up to the knee.

Roentgenograms showed pronounced osteo-articular destruction coupled with bony reorganization and the characteristic picture of metatarsal tapering, which has been described as resembling a "sucked candy-stick" (*sucre d'orge sucé*) (Fig. 6).

CASE 4.—Miss L.C., aged 35, has had diabetes for 25 years. She was admitted because of a tenosynovitis of the second finger of the right hand with resulting flexion deformity of the second, third and fourth fingers. She had been on a diet and has used insulin since her diabetes was recognized. She complained of decreased vision in the right eye. The thyroid gland was enlarged.

In her family history, it was noted that her mother and a maternal uncle were diabetic. Her personal history included an appendectomy in 1955. Nephritis occurred during an episode of diabetic acidosis in 1944. Many joints have shown signs of active rheumatoid arthritis since 1961. She also had Dupuytren's contractures in both hands. Moderate anemia was also noted.

The patient had noticed painless edema of her left leg and ankle. The heart and lungs were clinically normal on admission. The abdomen was soft. The liver was not enlarged, but there was moderate splenomegaly. Blood pressure was 170/80 mm. Hg. Knee and ankle jerks were absent bilaterally. The pupils reacted sluggishly to light. Pulses were felt in the feet, which were warm. This patient was thought to have developed the Kimmelstiel-Wilson syndrome as a complication of her diabetes. Perception of light touch was diminished in the lower third of her left leg and foot. There was also absence of position sense in her left great toe.

An appreciable normochromic anemia was present; since it occurred in combination with renal insufficiency, hypoplasia of the marrow was the most likely cause.

The pupils were equal, reacting sluggishly to light but with a slower reaction on the left. There was no Argyll Robertson pupil. Ocular tension was normal. There was early proliferative retinitis in the right eye with occlusion of a lower branch of the central vein and spreading hemorrhages. There were many punctate hemorrhages in the left eye, many exudates, and slightly enlarged veins without arteriolar nicking. In fact, proliferative retinitis was present in the right eye and diabetic retinopathy in the left.

The cardiologist described a pericardial friction rub, considered to be of renal origin. At that time, the patient was in the azotemic stage of her glomerulosclerosis.

Radiography of the left ankle showed an important area of bony destruction of the proximal part of the talus affecting also the antero-superior articular facet of the calcaneus, as well as the anterior facet of the tarsal scaphoid (Fig. 7). These areas of bony destruction were well localized and of pseudo-lacunar appearance; vascular calcification was evident.

Table I gives a résumé of the laboratory reports on these four cases.

DISCUSSION

The first case of diabetic neuroarthropathy in the American literature was reported in 1936 by Jordan.[35] He considered the lesion to be a diabetic process of neurotrophic nature. In 1947, Bailey and Root[6] found 17 cases after reviewing the records of 20,000 diabetics, a frequency of 1 per 1100. In 1953, Martin[44] reported nine cases among 150 patients with diabetic neuropathy. The latest important series of diabetic neuroarthropathies was published by Miller and Lichtman[46] in 1955; they

described the etiology in 31 cases of various neuroarthropathies of the foot, as follows: diabetes 17, tabes 4, myelodysplasia 4, idiopathic 2, alcoholism 1, medullary trauma 1, and leprosy 2.

In the 17 cases of diabetic arthropathy of the foot, the joints involved were as follows: tibiotarsal 1, at the tarsus 1, tarso-metatarsal 9, metatarso-phalangeal 8, and at the interphalangeal 2. The average age of these 17 patients was 53.17 years. Boehm[12a] notes an almost even distribution between males and females. This complication appears between 23 and 69 years of age, but the average patient is most commonly in the sixties.

TABLE II.—DIABETIC NEUROARTHROPATHIES OF THE FOOT (91 CASES)

Authors	*Date*	*Observations*
Jordan[35]	1936	1
Dreyfus and Zarachovitch[28]	1937	1
Bailey and Root[5]	1942	2
Jordan[36]	1943	2
Bailey and Root[6]	1947	17
Foster and Bassett[26]	1947	2
Morris[47]	1947	1
Muri[48]	1949	1
Wilson, McIntyre and Albertson[60]	1949	1
Lister and Maudsley[41]	1951	1
Parsons and Norton[50]	1951	2
Beidleman and Duncan[8]	1952	4
Martin[44]	1953	9
Azerad[3]	1953	2
Bénard *et al.*[9]	1953	1
Cram[19]	1953	1
Paul[51]	1953	2
Sheppe[55]	1953	1
Antes[2]	1954	1
Boulet, Mirouze and Pélissier[13]	1954	1
Miller and Lichtman[46]	1955	17
Lippman and Grow[40]	1955	2
Bolen[12b]	1956	2
Jacobs[33]	1958	2
Aagenaes and Haagensen[1]	1959	2
Bloch-Michel, Cauchoix and Cambier[10,11]	1959	1
Petersen[52]	1960	1
Degenhardt and Goodwin[21]	1960	2
Boehm[12a]	1962	1
Azerad *et al.*[4]	1963	4
Robillard and Gagnon	1964	4

On the whole, we found about 100 case reports of diabetic arthropathy in the literature, of which 91 were in the foot (Table II). These are mostly scattered reports of one or two cases. Six were reported in the French literature by Dreyfus and Zarachovitch[28] in 1937; Azerad[3] in 1953; Bénard *et al.*[9] in 1953; Boulet, Mirouze and Pélissier[13] in 1954; Bloch-Michel, Cauchoix and Cambier[10, 11] in 1959; and Azerad *et al.*[4] in 1963. We also noted two reports in the Scandinavian literature by Haagensen and Aagenaes[30] in 1959 and Petersen[52] in 1960.

In addition we found 18 cases of diabetic neuroarthropathy other than of the foot (Table III), 12 involving the ankle, 5 the knee, and the only case involving the lumbar spine—that of Zucker and Marder[61] in 1952, which was proved at autopsy.

We looked for cases of neuroarthropathy at the Hôpital du Sacré-Cœur de Montréal from 1952 to 1962 inclusive (Table IV). Out of a total of 1639 diabetics hospitalized during this decade, we

TABLE III.—DIABETIC NEUROARTHROPATHIES AT SITES OTHER THAN THE FOOT (18 CASES)

Ankle	12 cases
Knee	5 "
Spine	1 case
Total	18 cases

Authors	*Years*	*Location*
Jordan[35]	1936	Ankle and foot
de Takats[22]	1945	Knee
Foster and Basset[26] (Case 2)	1947	Ankle and foot
Shore[56]	1947	Knee
Spear[57]	1947	Knee
Knutsson[38] (four cases)	1951	Ankle
Zucker and Marder[61]	1952	Lumbar spine
Cram[20] (Case 2)	1955	Ankle and knee
Miller and Lichtman[46]	1955	Ankle and foot
Bolen[12b] (Case 2)	1956	Ankle and foot
Jacobs[33] (Case 3)	1958	Ankle
Bloch-Michel, Cauchoix, Cambier[10,11]	1959	Ankle and foot
Petersen[52] (two cases)	1959	Ankle and knee

reviewed 85 records containing skeletal x-rays. With respect to the lower limbs, we reviewed 14 radiographs of the knees, five of the ankles and three of the feet, making a total of 22 radiographs. We did not find a single case of diabetic neuroarthropathy. No case has been published in the French-Canadian or English-Canadian medical literature from 1900 to 1963.

TABLE IV.—DIABETIC NEUROARTHROPATHIES AT THE HÔPITAL DU SACRÉ-CŒUR DE MONTRÉAL FROM 1952 TO 1962 INCLUSIVE

Total number of hospitalized diabetics	1639 patients	
Total number of files with skeletal x-rays reviewed	85 files	
Number of radiographs of lower limbs reviewed	Knees	14
	ankles	5
	feet	3
	Total	22

Results = No case of neuroarthropathy found.

N.B.: No case was found in French-Canadian medical literature (*Union Médicale du Canada* from 1900 to 1963).

The clinical picture is usually that of long-standing and poorly controlled diabetes (Table V). The foot loses its shape progressively, becomes shorter and wider, and collapses in plano-valgus, or it becomes convex, the so-called "rocker-bottom foot". This pseudo-tabetic (Charcot's cube-shaped) foot often shows chronic plantar ulceration. There is no true infection, but accompanying the marked edema there may be redness on the dorsum of the foot. The peripheral circulation is usually good. What is most striking about these patients is the total absence of pain or very slight pain experi-

TABLE V.—DIABETIC NEUROARTHROPATHIES: CLINICAL FEATURES

1. Long-standing diabetes, poorly controlled.
2. Short, wide, plano-valgus foot.
3. Plantar ulcers without symptoms of infection.
4. Good peripheral circulation.
5. Painless.
6. Peripheral neuropathy: disturbances of position and vibratory sense. Diminished or abolished reflexes.

enced when walking on this deformed foot. Finally, these patients show, to varying degrees, definite signs of diabetic peripheral neuropathy. In some cases one notes a diminution in or loss of the position or vibration sense, and invariably there is decrease or absence of the knee and ankle jerks. Vasomotor disturbances were studied by Foster and Bassett[26] and by Lippman and Grow.[40] In both groups the vasomotor system showed evidence of a syndrome of complete sympathetic denervation.

TABLE VI.—DIABETIC NEUROARTHROPATHIES: RADIOLOGICAL FEATURES

1. Osteolytic lesions: Lysis, fragmentation, osteoarticular destruction.
2. Periostal reactions: New bone formation, osteophytes.

Radiological findings resemble closely those in any Charcot's arthropathy[17] (Table VI): there are areas of bony lysis with fragmentation, joint-space obliteration and, later, marked destruction of the osteoarticular structure. The picture of metatarsal tapering or "sucked candy-stick" osteolysis is very common. At times one can see an apparent bursting of the first metatarsal head. Moreover, certain areas show periosteal apposition with osteophytosis simulating peridiaphyseal "drippings".

The radiographs are so suggestive of tumours or osteomyelitis that amputations or occasional biopsies had been done and were available for a study of the histopathology in a few specimens. These biopsy specimens showed a loss of the general bony structure with many spicules in a fibrous matrix. The fibrous tissue contained focal or diffuse infiltration of plasmacytes, lymphocytes and histiocytes. Some long spicules adjacent to the periosteum were fringed by osteoblasts, but more often an osteoclastic activity was noticed. A fibroblastic proliferation was obvious and some arterioles showed a thickening of their intima (Table VII).

TABLE VII.—DIABETIC NEUROARTHROPATHIES. HISTOPATHOLOGY

1. Fibrous matrix containing bony spicules, plasmacytes, lymphocytes and histiocytes.
2. Osteoblastic and osteoclastic activity bordering the spicules.
3. Fibroblastic proliferation and thickening of arteriolar intima.

The only alterations occasionally found in laboratory study were secondary anemia and a moderate increase in the proteins of the cerebrospinal fluid, in the presence of normal pressure and cytology.

TABLE VIII.—DIABETIC NEUROARTHROPATHIES. DIFFERENTIAL DIAGNOSIS

1. Diabetes.
2. Syphilis or tabes.
3. Syringomyelia, mutilating acropathies.
4. Myelodysplasia: rachischisis, spina bifida.
5. Neuropathic leprosy.
6. Poliomyelitis.
7. Peripheral nerve lesions.
8. Alcoholism.
9. Tumours.
10. Pyogenic or tuberculous osteitis.
11. Congenital indifference to pain.
12. Arthropathy due to intra-articular corticosteroid injection.

In a patient with neuroarthropathy, the differential diagnosis must include diabetes, syphilis, neuropathic leprosy and, finally, poliomyelitis (Table VIII). Moreover, peripheral nerve lesions, alcoholism, tumours and pyogenic or tuberculous osteitis may show a clinical and radiological picture very similar to neurogenic arthropathies.

In syringomyelia, as in other mutilating acropathies, osteoarticular destruction will usually and progressively affect all four limbs (Fig. 8). Here,

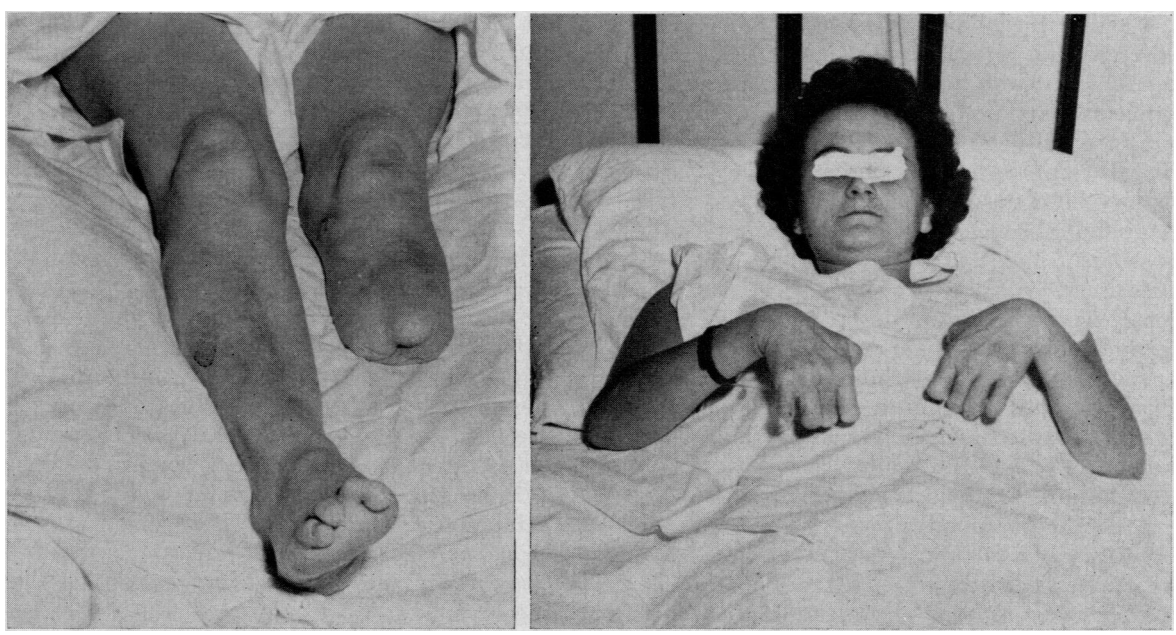

Fig. 8.—Mutilating disease of hands and feet.

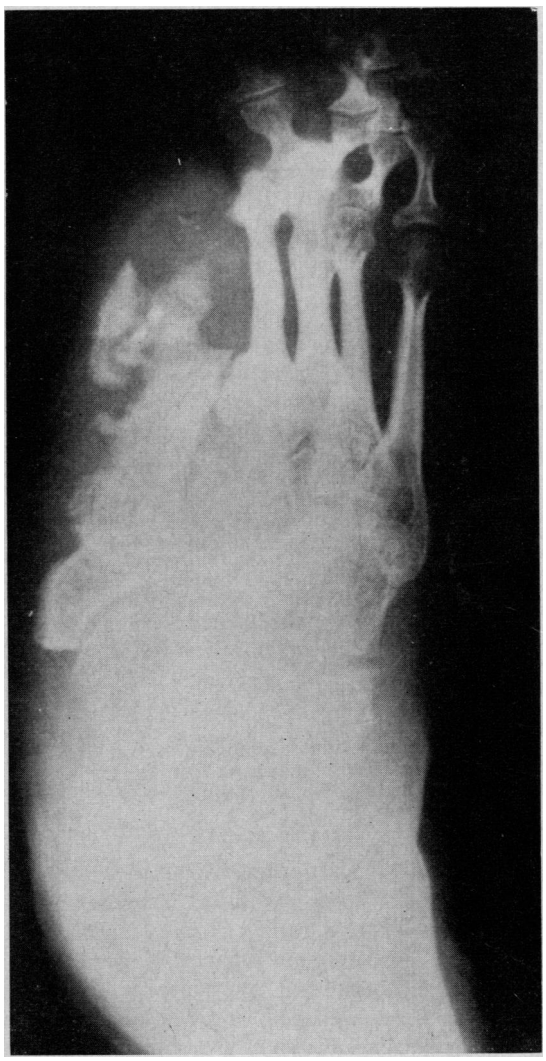

Fig. 9.—Radiological evidence of mutilation of digits.

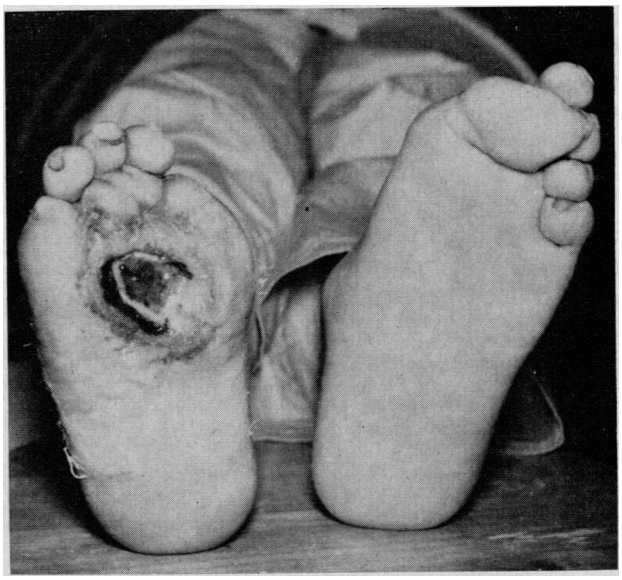

Fig. 10.—Chronic plantar ulceration associated with rachischisis: clinical appearance.

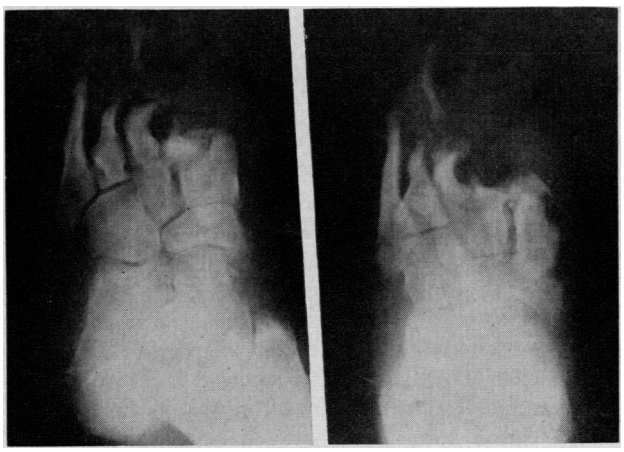

Fig. 11.—Osteoarticular destruction in same patient (Fig. 10).

radiographs are identical to those taken of the tabetic foot or any other Charcot's arthropathy (Fig. 9). Chronic plantar ulcerations occur in such myelodysplasias as rachischisis and spina bifida (Fig. 10). In these conditions, osteoarticular destruction is similar to that seen in a Charcot's tabetic foot (Fig. 11).

It is also appropriate to mention that Charcot's joints have been described in the unusual syndrome of congenital absence of pain sense. A few of these observations have appeared during the past decade.[7, 15, 18, 24, 25, 49, 53, 59] In 1938, Ford and Wilkins[25] were the first to describe this rare entity. Congenital absence of pain sense appears at birth, is permanent, covers the whole body and includes all forms of painful stimuli: pricking, burns, intense cold, faradic shock, and even the physiological phenomena which normally accompany those painful stimuli, such as acceleration of the pulse and elevation of blood pressure. These patients end with painless joint destruction typical of Charcot's neuroarthropathy.

Finally, some authors[16] have reported rapid and painless joint destruction following intra-articular injections of corticosteroid for degenerative arthritis. These arthropathies are clinically and radiologically similar to all other Charcot's neuroarthropathies. They appear to be the result of three associated factors: pain relief resulting from corticosteroid therapy, increased use of the damaged joint and, finally, the focal osteoporosis brought about by a large dose of corticosteroid.

Recently, Boehm[12a] summarized the etiologic and pathologic aspects of diabetic neuroarthropathies. Hodgson, Pugh and Young,[32] starting from the radiological similarity between diabetic arthropathy and the lesions of chronic osteomyelitis, suggested that infection was an etiological agent. There has been no actual proof to support this assertion. In 1951, Parsons and Norton[50] suggested that ischemia was the cause. However, the majority of these patients have normal pedal pulses, and oscillometric study usually reveals a perfectly normal circulation in both feet.

In 1947, Foster and Bassett[26] suggested that the causative lesion was in the autonomic nervous system. According to these authors, the patho-

genesis of this arthropathy appears to take place mostly in extramedullary nervous tissue, with the greatest damage in the posterior nerve roots, the spinal ganglia and the sensory fibres of peripheral nerves. It appears that the anterior roots, as well as the motor fibres of the peripheral nerves, are relatively spared. However, five years later, Martin[43] showed that every diabetic with neuropathy also has involvement of the autonomic nervous system affecting the feet. Therefore, because every patient with diabetic neuropathy does not have neuroarthropathy, this theory should be discounted. Tamlyn[58] has proved that Charcot joints are found in many diseases: syphilis, syringomyelia, myelodysplasia, and neuropathic leprosy as well as in diabetes mellitus. Eloesser[23] has shown that any joint deprived of sensation requires in addition some trauma to produce a typical Charcot joint. The common denominator in all these lesions is the lack of sensation; that is, any trauma, be it a single major injury or even repeated minor insults, can, in a denervated joint, cause neurogenic arthropathy.[31]

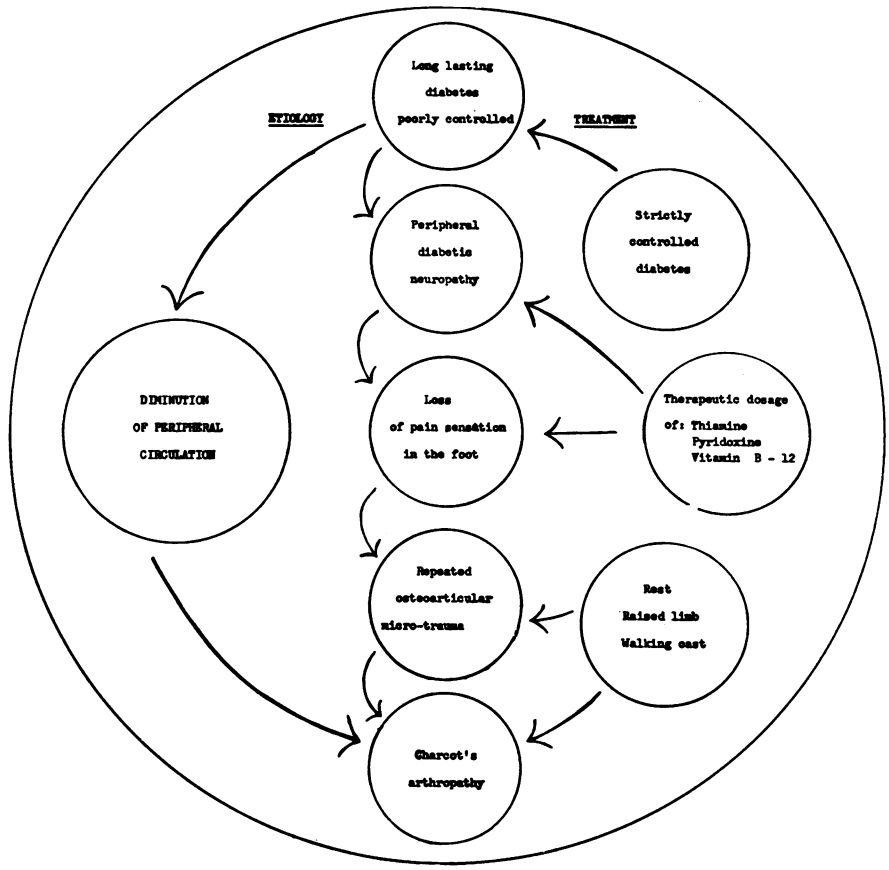

Fig. 12.—Diabetic neuroarthropathy. Etiology and treatment.

In the treatment of the neuropathy complicating diabetes, the use of thiamine, pyridoxine and vitamin B_{12} has often been recommended (Fig. 12). Lippman and Grow[40] appear to have produced a decrease in leg pain and promoted healing of pathological fractures by adrenocorticotrophic hormone (ACTH) administration. Degenhardt and Goodwin[21] recommended physiotherapy with faradic stimulation. Earlier authors tried plantar supports, laced boots and braces in an attempt to get an even weight distribution on the sole of the foot.[37] Lumbar sympathectomy was tried and initial results seemed promising, but other authors have rejected this operation for neuroarthropathies. Johnson[34] tried arthrodesis, but it is a technically difficult operation and the results were disappointing. In many cases the authors were so pessimistic that they recommended amputation.

The first therapeutic successes mentioned in the literature were those of Antes[2] and Boehm,[12a] who first raised the limb to reduce the edema and then applied a walking-cast to diminish pressure and trauma. In both cases, the patients showed a significant clinical and radiological improvement with bone re-formation. Antes' patient finally returned to his regular occupation six days a week.

The neurological syndrome recently stressed by Dreyfus, Zara and Cohen[29] belongs within the framework of sensory diabetic neuropathies, under the sub-heading "pseudo-tabetic type". To this group one must contrast the amyotrophic forms described by Garland[27] with which an arthropathy is never associated. Martin[45] has suggested that all diabetic patients with neuropathy should undergo routine radiographic studies of the feet.

Diabetic neuroarthropathy is still a relatively rare syndrome, but we believe that if greater attention is paid to this entity, observations similar to those contained in this communication might be made.

SUMMARY

Four typical cases of diabetic neuroarthropathy of the foot are described. Approximately 100 cases have been reported in the literature since 1936, of which 91 involved the foot and 18 the knee, ankle and lumbar spine. No such case was found in the files of the Hôpital du Sacré-Cœur de Montréal in the past 10 years. The authors found four cases, however, in a six-month period.

The clinical, radiological and histopathological features of this complication of diabetes are discussed.

Treatment consists of controlling the diabetes, treating the associated neuropathy and applying a walking-cast for several months.

RÉSUMÉ

Nous présentons quatre cas typiques de neuroarthropathie diabétique du pied. La revue de la littérature démontre qu'une centaine de cas ont été rapportés depuis 1936, dont 91 cas au pied et 18 cas au genou, à la cheville et au rachis lombaire. Aucun cas a été retrouvé à l'Hôpital du Sacré-Cœur de Montréal, au cours des dix dernières années, mais par contre, nous avons relevé quatre observations dans les six derniers mois.

Le tableau clinique, radiologique et histo-pathologique de cette entité est expliqué de même que le diagnostic différentiel et l'étiopathogénie de cette complication du diabète.

Le traitement consiste à équilibrer le diabète, à traiter la neuropathie associée et à appliquer un plâtre de marche pendant plusieurs mois.

We are grateful to the Records Department of the Hôpital du Sacré-Cœur de Montréal, and especially to Miss R. Chevalier for typing the manuscript. We also wish to thank our librarian, Miss C. Desserres, for the research work on the bibliography, and our photographer, Mr. Ferdinand Clark, for the illustrations. We appreciate the co-operation of Hoechst Pharmaceuticals for assistance in preparing the manuscript in English.

REFERENCES

1. AAGENAES, O. AND HAAGENSEN, N. R.: *Ugeskr. Leag.*, **121**: 572, 1959.
2. ANTES, E. H.: *J. A. M. A.*, **156**: 602, 1954.
3. AZERAD, E.: *Bull. et Mém. Soc. Méd. Hôp. Paris*, **69**: 302, 1953.
4. AZERAD, E. *et al.*: *Ann. Radiol. (Paris)*, **6**: 421, 1963.
5. BAILEY, C. C. AND ROOT, H. F.: *J. Clin. Invest.*, **21**: 649, 1942.
6. *Idem*: *New Eng. J. Med.*, **236**: 397, 1947.
7. BAXTER, D. W. AND OLSWESKI, J.: *Brain*, **83**: 381, 1960.
8. BEIDLEMAN, B. AND DUNCAN, G. G.: *Amer. J. Med.*, **12**: 43, 1952.
9. BÉNARD, H. *et al.*: *Sem. Hôp. Paris*, **29**: 3487, 1953.
10. BLOCH-MICHEL, H., CAUCHOIX, J. AND CAMBIER, J.: *Presse Méd.*, **67**: 809, 1959.
11. *Idem*: *Ibid.*, **67**: 842, 1959.
12a. BOEHM, H. J., JR.: *New Eng. J. Med.*, **267**: 185, 1962.
12b. BOLEN, J. G.: *Radiology*, **67**: 95, 1956.
13. BOULET, P., MIROUZE, J. AND PÉLISSIER, M.: *Sem. Hôp. Paris*, **30**: 2410, 1954.
14. BROCH, O. J. AND KLOVSTAD, O.: *Acta Med. Scand.*, **127**: 514, 1947.
15. CAMBIER, J. AND LEFEBVRE, J.-P.: *Presse Méd.*, **67**: 1551, 1959.
16. CHANDLER, G. N. *et al.*: *Brit. Med. J.*, **1**: 952, 1959.
17. CHARCOT, J.-M.: *Archives de Physiologie normale et pathologique*, **1**: 161, 1868.
18. COHEN OF BIRKENHEAD, LORD: *J. Bone Joint Surg.* [*Brit.*], **43-B**: 219, 1961.
19. CRAM, R. H.: *Surg. Clin. N. Amer.*, **33**: 1759, 1953.
20. *Idem*: *J. Bone Joint Surg.* [*Amer.*], **37-A**: 967, 1955.
21. DEGENHARDT, D. P. AND GOODWIN, M. A.: *J. Bone Joint Surg.* [*Brit.*], **42-B**: 769, 1960.
22. DE TAKATS, G.: *Proceedings of the American Diabetes Association*, **5**: 183, 1945.
23. ELOESSER, L.: *Ann. Surg.*, **66**: 201, 1917.
24. FEINDEL, W.: *J. Bone Joint Surg.* [*Brit.*], **35-B**: 402, 1953.
25. FORD, F. R. AND WILKINS, L.: *Bull. Hopkins Hosp.*, **62**: 448, 1938.
26. FOSTER, D. B. AND BASSETT, R. C.: *Arch. Neurol. Psychiat. (Chicago)*, **57**: 173, 1947.
27. GARLAND, H.: Diabetic amyotrophy. *In*: Modern trends in neurology, 2nd ed., edited by D. Williams, Butterworth & Co., Ltd., London, 1957, p. 229.
28. DREYFUS, G. AND ZARACHOVITCH: *Bull. et Mém. Soc. Méd. Hôp. Paris*, **53**: 328, 1937.
29. DREYFUS, G., ZARA, M. AND COHEN, A.: *Rev. Lyon. Méd.*, **7**: 561, 1958.
30. HAAGENSEN, N. R. AND AAGENAES, O.: *Ugeskr. Laeg.*, **121**: 569, 1959.
31. HALDEMAN, K. O. AND SOTO-HALL, R.: *J. A. M. A.*, **111**: 2043, 1938.
32. HODGSON, J. R., PUGH, D. G. AND YOUNG, H. H.: *Radiology*, **50**: 65, 1948.
33. JACOBS, J. E.: *J. Bone Joint Surg.* [*Amer.*], **40-A**: 1043, 1958.
34. JOHNSON, E. W., JR.: *W. Virginia Med. J.*, **54**: 157, 1958.
35. JORDAN, W. R.: *Arch. Intern. Med. (Chicago)*, **57**: 307, 1936.
36. *Idem*: *Southern Med. J.*, **36**: 45, 1943.
37. JOSLIN, E. P.: Unusual manifestations of diabetic neuropathy. Part 1. Charcot joints. *In*: Treatment of diabetes mellitus, 9th ed., edited by E. P. Joslin *et al.*, Lea & Febiger, Philadelphia, 1952, p. 484.
38. KNUTSSON, F.: *Acta Radiol. (Stockholm)*, **36**: 114, 1951.
39. KRAUS, W. M.: *Arch. Neurol. Psychiat. (Chicago)*, **7**: 202, 1922.
40. LIPPMAN, E. M. AND GROW, J. L.: *J. Bone Joint Surg.* [*Amer.*], **37-A**: 971, 1955.
41. LISTER, J. AND MAUDSLEY, R. H.: *Lancet*, **2**: 1110, 1951.
42. MARBLE, A.: *J. Kansas Med. Soc.*, **49**: 189, 1948.
43. MARTIN, M. M.: *Proc. Roy. Soc. Med.*, **45**: 503, 1952.
44. *Idem*: *Brain*, **76**: 594, 1953.
45. *Idem*: *Proc. Roy. Soc. Med.*, **47**: 139, 1954.
46. MILLER, D. S. AND LICHTMAN, W. F.: *A.M.A. Arch. Surg.*, **70**: 513, 1955.
47. MORRIS, M. H.: *New York J. Med.*, **47**: 1395, 1947.
48. MURI, J.: *Acta Med. Scand.*, **135**: 391, 1949.
49. MURRAY, R. O.: *Brit. J. Radiol.*, **30**: 2, 1957.
50. PARSONS, H. AND NORTON, W. S., II: *New Eng. J. Med.*, **244**: 935, 1951.
51. PAUL, J. T.: *Amer. Pract. Digest Treat.*, **4**: 49, 1953.
52. PETERSEN, A.: *Acta Orthop. Scand.*, **30**: 217, 1960.
53. PETRIE, J. G.: *J. Bone Joint Surg.* [*Brit.*], **35-B**: 399, 1953.
54. RUNDLES, R. W.: *Medicine (Balt.)*, **24**: 111, 1945.
55. SHEPPE, W. M.: *Ann. Intern. Med.*, **39**: 625, 1953.
56. SHORE, T. H. G.: *Lancet*, **2**: 738, 1947.
57. SPEAR, G. E.: *Ibid.*, **2**: 963, 1947.
58. TAMLYN, T. T.: *Minnesota Med.*, **40**: 117, 1957.
59. VAN DER HOUWEN, H.: *J. Bone Joint Surg.* [*Brit.*], **43-B**: 314, 1961.
60. WILSON, I. H., MCINTYRE, C. H. AND ALBERTSON, H. K.: *Calif. Med.*, **70**: 420, 1949.
61. ZUCKER, G. AND MARDER, M. J.: *Amer. J. Med.*, **12**: 118, 1952.

PAGES OUT OF THE PAST: FROM THE JOURNAL OF FIFTY YEARS AGO

ROBERT BARANY AND THE CALORIC REACTION

Although a considerable amount of experimental and practical work was being done by men like the physiologist Ewald, of Strassburg, the American psychologist, James, and Jansen, the Berlin otologist, who was the first to operate on suppurative diseases of the semicircular canals, still the clinical side remained undeveloped. This was mainly due to the fact that there was no method at their command by which they could investigate a unilateral affection of the semicircular canals, because the turning test, which was the one employed, irritated the semicircular canals on both sides, and the galvanic experiment gave uncertain and unreliable results, as it still does to-day.

All this was changed when Robert Bárány made his epoch-making discovery of the caloric reaction in 1905. The history of it is quite interesting. As a young assistant of Politzer's in the Allgemeine Krankenhaus in Vienna, Bárány was one day syringing an ear with very cold water, when the patient began complaining of dizziness, and on observing the eyes, he noticed there was a combined rotary and horizontal nystagmus away from the syringed ear. Thinking that the extreme temperature of the water was possibly responsible for the dizziness, he ordered warmer water and this time the water was very hot. He syringed the same ear, the patient made similar complaints, and again observing the eyes, he noticed much to his surprise, an identical movement but toward the irrigated ear. He then syringed the ear with water of body temperature and obtained no reaction. At this moment, he recognized that it must be the temperature of the water which is responsible for the dizziness, and for the nystagmus—its character and direction.

So he proved, after examining hundreds of individuals of all ages, with normal and destroyed drums, but intact labyrinths, that the normal caloric reaction was obtainable in all these cases, i.e. in syringing an ear with water of body temperature, no nystagmus is produced, nor any other ill effects, as dizziness, disturbance of equilibrium, nausea or vomiting, while cold water produces in a short time, a nystagmus in the direction away from the irrigated ear, and hot water, similar nystagmus, but towards the same side. He further studied cases with pathological conditions of the internal ear and found that where the semicircular canals or vestibular nerve were destroyed by cholesteatoma, labyrinthitis, tumour, etc., the caloric reaction was absent on that side. Operation or postmortem examination confirmed these findings.—D. H. Ballon, *Canad. Med. Ass. J.*, 4: 872, 1914.